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Dandy-Walker Syndrome

— Report of Two Cases —

by

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Hydrocephalus caused by congenital atresia of the foramina of LUSCHKA and MAGENDIE was first reported by DANDY and BLACKFAN[2] in 1914. DANDY[3] described later its diagnosis and treatment. Similar cases were reported by TAGGART and WALKER[7] in 1942 and by WALKER[8] in 1944. They concluded that the cause of the typical deformity was failure of the foramen of MAGENDIE to open during intrauterine life.

Although these authors threw a light upon the approach to the pathogenesis of congenital hydrocephalus, this pathological entity was overlooked by neurosurgeons until BENDA[1] suggested to call it as "DANDY-WALKER syndrome" on the basis of detailed neuropathological observations in 1954. According to his definition, DANDY-WALKER syndrome is not only a sole occlusion of the foramen MAGENDIE and LUSCHKAE, but cystic dilatation of the fourth ventricle with hypoplastic cerebellar vermis.

Recently we experienced two cases of this specific type of hydrocephalus, one of which was thought to be the severe infantile form and the other atypical one.

Case 1. H.K. A 7-month-old male baby was admitted on December 7, 1965 with an enlarged head and vomiting. He had been delivered as a matured neonate and grown up normally until his parents noticed the excessive growth of his head two months after birth. He had been admitted to the pediatric department in this hospital for a month at his age of the third to fourth month, and diagnosed as hydrocephalus and congenital heart failure. Several weeks prior to admission, vomiting and transient attacks of cyanosis had begun to occur.

On admission, he was looked as a pale emaciated baby with enormously large dolichocephalic head. His eyes were exophthalmic and turned to downward. Circumference of the head was measured as 58 cm. Fontanelle was dilated measuring as 19×8 cm. Skin over the head was tense and glossy with dilatation of the veins. Cracked pot sound was heard on percussion of the head. Congenital deformity was found in the lobule of his right ear. Respiration was regular but somewhat superficial. Systoric murmur was heard everywhere in his chest.

On examination, there were no neurological abnormalities. Plain skull x-ray revealed very thin skull especially at the prominent occiput. Lumbar puncture was not done, and emergency ventriculostomy was performed on the day of admission. Pressure of ventricular fluid was not measured but it seemed to be high since it squirted out of the needle. Fine polyethylene tube was inserted into the dilated lateral ventricle connecting to the pressure

regulator. Ventricular pressure was maintained at 150 mm H_2O.

After the operation, vomiting ceased and he looked like to regain his vitality in crying and moving. The size of the head became smaller and fontanelle depressed. Venous dilatation over the head became obscure. He went so well during the first postoperative week that we were planning to perform the ventriculoatrial shunt in the next week.

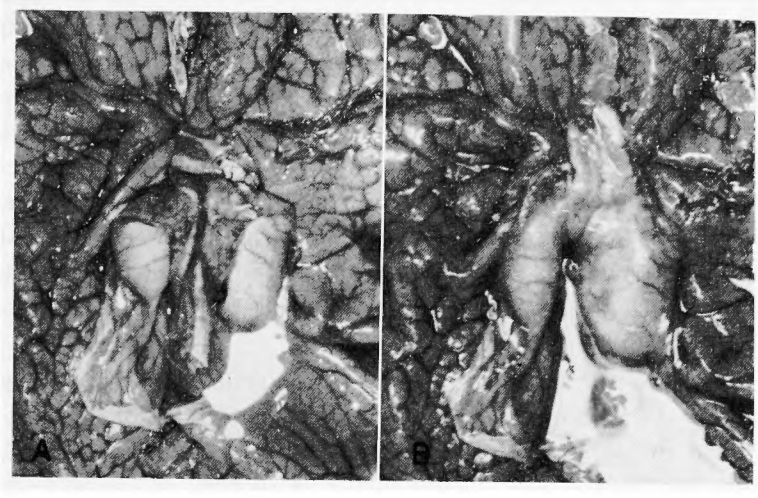

Fig 1 Basal view of the fresh brain of case 1.
(A) Extraordinally hypoplastic cerebellar hemispheres are completely divided bilaterally. Cerebellar tonsils and vermis are absent. Note the cystic membrane attached to the dorsolateral margin of the cerebellum. (B) When flattened and flexible brain stem is turned over, outlet of the aqeduct is directly seen through the unroofed fourth ventricle.

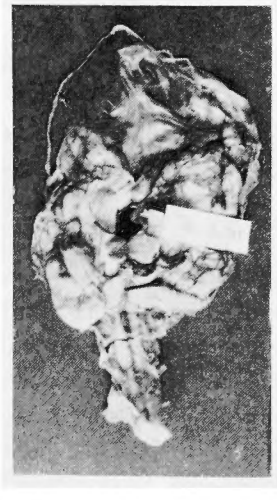

Fig. 2 Formalin fixed hind-brain structures (case 1). Partly thickened cyst membrane is attached to the dorsolateral margin of the flattened cerebellar hemispheres.

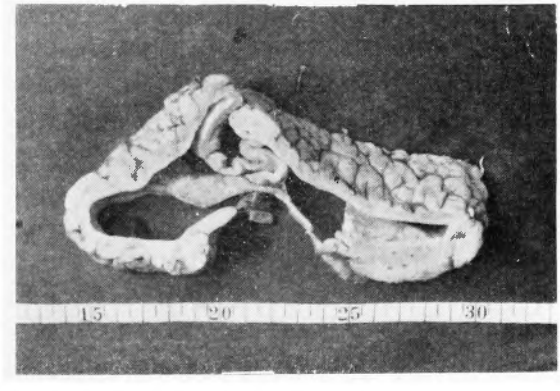

Fig. 3 Frontal section of the cerebrum of case 1 (fixed in formalin). Lateral ventricles are enormously dilated. Basal part of the temporal lobe is extremely thinned out.

However, he resumed to have vomiting at the end of the first week in spite of low ventricular pressure. He gradually became worse in the next week, and died on the tenth hospital day.

An autopsy revealed extremely dilated cerebral ventricles. The fresh specimen of the brain was hard to be held because of the thin cerebral cortex. Posterior fossa was markedly dilated and occupied by huge cyst encapsulated by thin fibrous membrane which were firmly attached to the lower margin of the fourth ventricle, extending on the surface of the cerebellum toward the tentorium and the region of the foramen magnum. Marked hypoplasia of the cerebellum with agenesis of the cerebellar vermis was noted. Brain stem structures were flattened probably due to compression by the cyst. When the flexible brain stem was turned over, the aqueduct of SYLVIUS was directly seen through the dilated fourth ventricle. (Figs. 1 and 2) Frontal section of the cerebrum showed enormously dilated lateral ventricles with cortical atrophy. Dilatation of the lateral ventricle was greater the left than the right, so that the basal face of the left temporal lobe appeared to be membraneous. (Fig. 3) The failure of closure of the foramen ovale was found in the heart.

Case 2. M.H. A 9-year old girl was first seen at the psychiatric department in this hospital on January 14, 1965, and then referred to our clinic on January 29 with a diagnosis of "brain tumor". Since a couple of month before hospitalization, her parents have noticed the sluggishness and idleness in her behavior.

On examination, the general physical findings were entirely normal. Higher mental functions were almost intact. Cranial nerves normal. No nystagmus. Mild unsteadiness on her feet and hyperactive knee and ankle jerks on both sides were noted. Extensor plantal responses, positive CHADOCK reflex and clonus were observed in her feet. Spinal fluid was under a pressure of 180 mm H_2O, clear fluid, no abnormal chemical properties, and normal QUECKENSTEDT's phenomenon.

Subsequent right carotid angiogram revealed no abnormalities. An EEG showed diffuse abnormal slow pattern with temporal desynchronization.

She had been under psychiatric care until she appeared again at our clinic on October 6. At this time she was moderately obtunded and aphasic. She had difficulty in swallowing and unable to stand on her feet. She was transferred to our clinic on the 12th of October, 1965.

On admission, she was stuporous and lying quietly on her back. Pupils were equal in size, both reacted directly and consensually to light. Conjugate deviation to the right with virtually continuous eye movements were present, and there were full up and down excursions on doll's maneuver. Corneal reflexes were present but equally decreased. No remarkable facial paresis. No response to loud sounds. She had some difficulty in swallowing and unable to eat, smack lips and yawn. Her right arm was spastic, pronated at elbow joint. Left arm was extended but slightly rigid on passive flexion. The deep tendon reflexes were present and equally depressed in both upper extremities. Occasionally moved all limbs spontaneously. The knee and ankle jerks were equally exaggerated and easily elicitable of the clonus. Toe responses were extensor. All limbs withdrew from painful stimuli.

Lumbar puncture yielded 5 ml. of clear acellular fluid under opening pressure of 130 mm H_2O and closing pressure 90 mm H_2O, with normal protein content. Carotid and vertebral arteriograms revealed no remarkable abnormalities. Pneumoencephalogram showed moderate dilatation of the both lateral and third ventricles. Myodil ventriculography indicated the filling defects in the anterior part of the third ventricle and around the fourth ventricle. (Fig. 4)

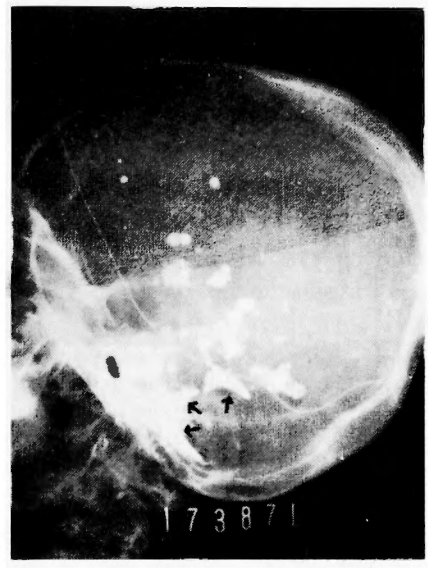

Fig. 4 Myodil ventriculagram of case 2. Hemispherical filling defect in the caudal part of the fourth ventricle is shown by arrows.

Right frontal craniotomy with exploration of the foramen of MONRO was performed on December 2nd, but no space occupying lesion in the third ventricle was found. Suboccipital craniectomy was carried out on the 24 th of February 1966. A pingpong-ball sized arachnoid cyst was protruded caudal from the fourth ventricle. Membraneous cyst wall was excised and suboccipital decompression was completed. The cerebellar tonsils were widely divided and the vermis of the cerebellum was absent.

Postoperatively, there was no definite improvement and no great changes in findings. Ventriculo-atrial shunt was intended on March 7 th, because of progressively increasing cerebrospinal fluid pressure. However, it was thought to be dangerous since the ventricular fluid was turbid at the time of operation. Instead of V-A shunt, lateral ventriculostomy with controlled cerebrospinal fluid drainage was performed. The sign of meningitis with high fever was developed postoperatively. She expired 5 days after the ventriculostomy.

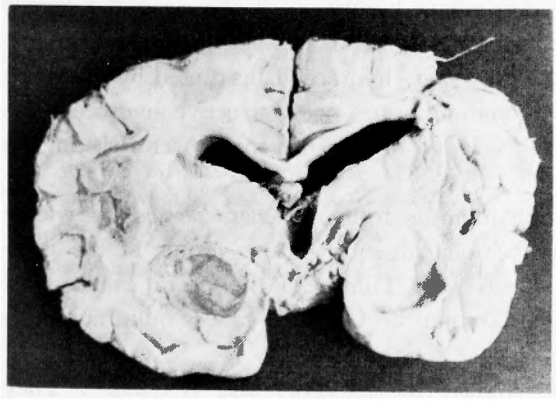

Fig. 5 Frontal section of the cerebrum of case 2 (fixed in formalin). Dilatation is marked in both temporal horns of the lateral ventricles and the third ventricle.

On autopsy, there were no remarkable changes in the cerebrum except for moderately dilated lateral and third ventricle containing suppurative cerebrospinal fluid. (Fig. 5) Cerebellar hemispheres were nearly intact. The most conspicuous abnormal findings were agenesis of the cerebellar vermis and atrophy of the pons.

The existence of the arachnoid cyst at the caudal part of the fourth ventricle was evident though the cystic membrane had been already removed at the operation. Foramina of LUSCHKA were seemed to be patent. (Fig. 6)

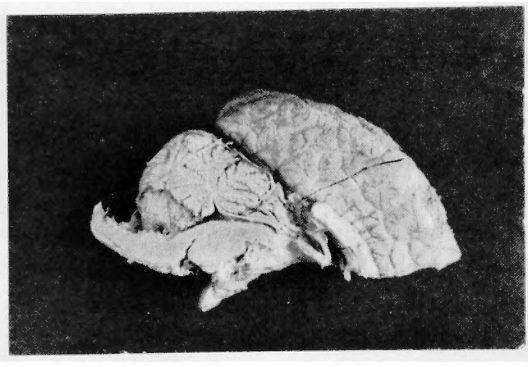

Fig. 6 Sagital midline section of the hind-brain structures and posterior part of the cerebrum (case 2). Indentation at the caudal part of the cerebellum represents the site of the cyst. Part of the cyst membrane is remained over the medulla. Atrophy of the pons is observed. Dilatation of the fourth ventricle is not evident.

DISCUSSION

Hydrocephalus is grossly classified into two groups ; communicating and non-communicating hydrocephalus, depending upon the communicability between the ventricles and the subarachnoid space. Clinically, the communicability is able to be estimated by testing whether dye passes from the ventricular system to the lumbar subarachnoid space or not.

Communicating hydrocephalus is caused by either excessive production of cerebrospinal fluid or disturbance in absorption of it. Sometimes, venous occlusion or arachnoidal adhesions are also responsible for the communicating hydrocephalus.

On the other hand, the cause of non-communicating hydrocephalus is varied mainly on the basis of the localization of the occlusion in the ventricular system. The foramen of MONRO, aqueduct of SYLVIUS, foramen of MAGENDIE and foramina of LUSCHKA are the main sites of occlusion, and they are apt to occlude during the embryonal life. DANDY-WALKER syndrome belongs to this category of hydrocephalus.

ARNOLD-CHIARI malformation, namely, descending herniation of the cerebellar tonsils through the foramen magnum into the spinal canal with or without meningomyelocele, is usually separated from above classification. However, hydrocephalus caused by ARNOLD-CHIARI malformation must belong to non-communicating or obstructive hydrocephalus because of the cerebrospinal fluid pathway is occluded around the fourth ventricle in this condition.

DANDY-WALKER syndrome, especially the infantile forms, is characterized by widely separated cerebellar hemispheres. There may be total absence of the vermis of the cerebellum, and there is a bulging posterior fossa with failure of the lateral sinuses and attached tentorium to descend from their fetal position.[3)7)] Our first case is quite agreeable with this criteria from the autopsy findings.

The pathogenesis of this condition was studied by TAGGART and WALKER[7)]. They concluded that the cause of the typical deformity was failure of the foramen of MAGENDIE to open during intrauterine life. They believed that the associated cerebellar lesions, although prominent, are the result of the hydrocephalus. On the other hand, BENDA[1)]

believed that the lesion belonged to the category of cleft formations or rachischisis, and that the essential fault was not in the failure of the apertures to open, but failure of the dorsal midline to fuse.

The most reasonable explanation of the pathogenesis of the DANDY-WALKER syndrome was made by GARDNER and his collaborates[4]. They suggested that the varied pathological entities which cause hydrocephalus such as ARNOLD-CHIARI malformation, DANDY-WALKER syndrome, and arachnoid cyst of the cerebellum are sole varying expressions of embryonal atresia of the fourth ventricle. According to their hydromyelic theory, shortly after the closure of the neural tube at the fifth embryonal week, the choroid plexus begins to pour fluid into this closed cavity and results in the enlargement of the cavity. This enlargement will be compensated by outflow of the fluid out of the cavity due to the increased permeability of the rhombic roof, and subsequent absorption into the subarachnoid space. The permeability of the membrane of the rhombic roof exerts an influence upon the form of above expressions. It is obvious that the less permeable the membrane, the severer will be the manifestations and the earlier will they appear. The form of the local expression, on the other hand, depends upon the degree of elasticity in the membrane. If it is more elastic it will stretch to form diverticulum of the fourth ventricle which may herniate through the foramen magnum and forms DANDY-WALKER syndrome. If inelastic, the hind-brain itself will herniate and forms ARNOLD-CHIARI malformation. If sufficiently fragile, it will rupture spontaneously and may this convert an obstructive into a communicating hydrocephalus. If the occluding membrane splits into two layers, fluid may collect between them and form so called arachnoid cyst of the cerebellum.

According to the GARDNER's opinion, our second case is to be expressed as a diverticulum of the framen of MAGENDIE which is synonymous of adult form of DANDY-WALKER syndrome.

GARDNER described the similar case in which ventricles had some communication with the subarachnoid space, in spite of the existence of the thick bulging membrane at the occluded foramen of MAGENDIE.

Explaining this particular case, he emphasized the importance of the patency of the midline foramen than patency of the laterally placed and devious foramens of LUSCHKA, because the foramen MAGENDIE is situated in a more advantageous position to permit a spinal subarachnoid dissipation of the ventricular fluid pulse wave as it spurts from the aqueduct with each heart beat[4].

Diagnosis of DANDY-WALKER syndrome is relatively easy in the severe infantile form. However, mild adult form is difficult to diagnose, which is often misdiagnosed as "arachnoid cyst simulating brain tumor". In non-communicating hydrocephalus, shape of the head is characteristic according to the site of occlusion. Enlargement of the head is outstanding at the occipital region in DANDY-WALKER syndrome, while in the case of the obstruction at the foramen of MONRO or aqueduct, enlargement of the forehead is prominent.

On skull x-rays in case of DANDY-WALKER syndrome the inion is elevated and the impressions of the transverse and straight sinuses are in the posterior parietal bones instead of being located in the occipital bones. In the AP projection, the bitemporal distance

appears less marked than in other types of hydrocephalus. The lateral projection reveals a thinning of the lower occipital bones, and the posterior cranial fossa appears markedly enlarged[6].

Pneumoencephalography reveals dilatation of the whole ventrciular system, especially characteristic feature of this condition is the dilatation of the aqueduct communicating with a large cyst-like formation in the posterior fossa[6]. MATSON[5] recommended the upside down position in air study. In this unusual position, the projection of the cyst of the posterior fossa into the upper cervical spinal canal may be seen.

The principle of the treatment of DANDY-WALKER syndrome consists of suboccipital craniectomy with excision of the cyst wall as widely as possible and shunting procedure for hydrocephalus. In cases of DANDY-WALKER syndrome with advanced hydrocephalus are most effectively treated by ventriculo-atrial shunt following the excision of the cyst wall[5].

Satisfactory surgical results can be expected if the lesion is properly treated in earlier stage.

SUMMARY

Two cases of DANDY-WALKER syndrome characterized by cystic dilatation of the fourth ventricle due to the atresia of the foramina MAGENDIE and LUSCHKA are reported. The first case is regarded as a severe infantile form and the second case belongs to a category of the diverticulum of the fourth ventricle or mild adult form.

The common findings of both cases in autopsy are the cyst formation of the caudal part of the fourth ventricle and agenesis of the cerebellar vermis, although hydrocephalus and maldevelopment of the cerebellum are much more prominent in the first case.

The pathogenesis, diagnosis and treatment of this malformation are briefly discussed.

This pathological condition would not be so rare as long as careful examination is pursued. And the improvement may be expected if suitable operative treatment is performed in earlier stage.

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和文抄録

Dandy-Walker 症候群の2例

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東　健一郎・森信昭二・望月一徳

第1例は7ヵ月の男児で著るしい水頭症と嘔吐を主訴として来院した．入院時は頻発する嘔吐のために栄養障害と脱水状態が著るしく，頭部は長頭形を呈し．頭囲は58cmで，大泉門の拡大・膨隆を認めた．脳室ドレナージにより一時症状は好転していたが，入院10日目に死亡した．剖検により後頭蓋窩は巨大な第4脳室の嚢胞によつて占められ，Magendie孔およびLuschka孔は閉塞し，小脳は著明に萎縮して母指頭大の半球を両側に認めるのみであつた．

第2例は9才の女児で入院時は昏睡状態であり，言語障害，嚥下障害，尿尿の失禁，両上肢の痙性麻痺，下肢の病的反射，右方への共同偏視などを認めた．髄液圧は低く，蛋白・糖・細胞数は正常で，気脳写により側脳室および第3脳室の中等度の拡大を認めた．マイオジールによる脳室写で第4脳室下半の半円形圧迫像を認めたので，後頭蓋窩開頭術を行なつたところ，Magendie 孔の閉塞と，この部分の嚢胞，および小脳虫部の欠損を認めた．術後症状の改善はみられず．化膿性髄膜炎を併発して死亡した．剖検によつて小脳虫部の形成不全と，圧迫によると思われる橋の萎縮を確認した．

この2症例に共通した所見は，Magendie 孔の閉塞による第4脳室の嚢胞性拡大と，小脳虫部の形成不全であつて，ともに Dandy-Walker 症候群の定義に一致する．第1例は典型的な重症小児型であるが，第2例は第4脳室憩室ともよばれている非典型的な遅発型であり，このような症例は早期に発見して手術的治療を行えば治癒も可能である．